

Autoimmune Hepatitis (AIH)

Leaflet for paediatric physicians

Autoimmune hepatitis (AIH) can affect children, even infants, and adolescents. Here we summarise to what extent children and adolescents with AIH differ from adult patients.

Disease presentation

- The presentation of autoimmune hepatitis (AIH) is often more aggressive than in adults, but also clinical courses with long-time subclinical AIH are common.
- Overlap between AIH, sclerosing cholangitis and IBD is more frequent in childhood
- There is a significant risk of relapse especially during adolescence due to hormonal changes or non-adherence
- Diagnostic autoantibody titres are often lower in paediatric AIH patients than in adults, and therefore the diagnostic criteria vary between adults and children
- The autoantibodies anti-LC1 and anti-LKM are more frequent in children and adolescents than in adults. The presence of anti-LC1 and/or anti-LKM define type 2 AIH.
- Genetic variations for disease risk and virus-infections, which may trigger AIH are different
- Liver biopsy shows a more mixed histological picture
- An overlap syndrome between AIH and sclerosing cholangitis (which is called 'autoimmune sclerosing cholangitis' (ASC)) occurs more often in children than adults. Every child diagnosed with AIH should undergo cholangiography (MRI/MRCP, Magnetic Resonance Imaging/Magnetic Resonance Cholangio-Pancreatography) in the diagnostic work-up
- Approximately one in five children with AIH have associated autoimmune diseases such as inflammatory bowel disease (IBD), celiac, thyroid or rheumatic disease
- Differential diagnoses, which must be considered, are different
- Children with AIH should be supported, like other children with chronic diseases, in their Transition-Process, this must be implemented in the medical care service, also to avoid the higher risk of non-adherence in adolescents.

Treatment

Paediatric AIH most often responds well to immunosuppressive therapy. However, the treatment of children with AIH must take other aspects such as growth and puberty into account. Under treatment, relapse of the disease can occur which is often due to non-adherence, particularly in adolescents. However, children with AIH type 2 are also in risk of relapse, despite good adherence. Psychoeducation of the child and teenager is part of the treatment. It is of uttermost importance that

the young person learns about the disease and understands the indication for the treatment, its side effects and the potential detrimental consequences of non-adherence as early as possible.

The side-effects of medication used to treat AIH may be different in children compared to those in adults.

There may be a risk that a paediatric AIH patient will need a liver transplant. The long-term risk factor will be the amount of scar tissues on the biopsy, the livers ability to regenerate during treatment and the overall response to treatment will influence the outcome.

Tests and investigations

Several tests can be indicated to diagnose and monitor paediatric AIH and screen for simultaneous diseases of other organs: blood tests, urine analysis, stool test (faecal calprotectin), abdominal ultrasound, magnetic resonance imaging to investigate the bile ducts (MRI/MRCP), liver biopsy, gastroduodenoscopy and/or colonoscopy. Liver biopsies and gastro-intestinal endoscopies are performed under deep sedation/general anaesthesia in children. MRI scans require sedation until the child can cooperate (normally around 6-7 years of age). As procedures can be a challenge, especially in young children, it is very important that both the caregivers and the child are well informed on what will happen.

The tools available in the paediatric department to minimise discomfort from procedures should be discussed with the patients and caregivers such as:

- Measures to numb the skin locally (EMLA®, cold spray)
- Tools to distract the child (TV, Virtual Reality glasses)
- Nitrous oxide
- Etc.

Here, we have listed some advice on how to minimise discomfort for the child. However, the parents know the child best and will know best how to support them.

- Before the procedure:
 - Give correct information on what will happen and tell what you have done to minimise discomfort.
 - Give the child the feeling that it can keep control on the situation e.g. let it decide if it wants to lie down, or sit on your lap, if it likes to watch its favourite movie or if its favourite bear should be there... However, never make the procedure itself be an option for the child.

- During the procedure:
 - Create a calm environment
 - If the situation turns out differently than previously explained, clarify why this happens
 - Simple relaxation techniques (such as taking deep slow breaths) and distraction can be helpful (e.g. tell me a story, should I tell you a story, what have you done in school today, singing).
 - Give clear information on the procedure avoiding words which might evoke anxiety (such as “pain”, needle, e.g. when I count to three you will have a warm feeling in your arm, that is when the nurse gives you the medicine).
- After the procedure:
 - Congratulate the child and reinforce the positive things they did during the procedure (you were brave, I think that it was great that you told me a story, it was difficult, and I am proud of you).
 - Document a plan for next time (what works, what should be different)

Vaccinations

- Provide data on the previous vaccination schedule of the child to the treating physician and discuss all future vaccinations with the treating physician.
- The response to vaccinations can be suppressed under high doses of immunosuppression. Therefore, it is advisable not to give scheduled vaccinations in the initial phase of the treatment or in the immediate phase after a relapse.
- Live attenuated vaccines have been contraindicated in patients taking immunosuppressive drugs, but recent development may change this into “relatively contraindicated”. Examples of live attenuated vaccines are: Varicella Zoster (chickenpox), Measles, Mumps and Rubella (MMR), yellow fever, BCG and oral typhoid vaccine.
- Dead vaccines can be given but the timing should be discussed with the treating physician. Examples of dead vaccines are: Pneumococcal, Haemophilus Influenza, Diphtheria, Tetanus, Pertussis (whooping cough), HPV vaccine.
- It is recommended to check Hepatitis B antibodies yearly and provide a booster vaccine if the antibodies turn out to be insufficient.
- Hepatitis A vaccination is recommended.
- Annual flu vaccination is recommended.